

Surgical outcomes of 380 patients with double outlet right ventricle who underwent biventricular repair

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Objectives: The study objective was to report the outcomes of biventricular repair in patients with double outlet right ventricle.

Methods: Patients with double outlet right ventricle who underwent biventricular repair at Fuwai Hospital from January 2005 to December 2012 were included. Patients were excluded if double outlet right ventricle was combined with atrioventricular septal defect, heterotaxy syndrome, atrioventricular discordance, or univentricular physiology.

Results: A total of 380 consecutive patients with a mean age of 1.9 ± 2.1 years (range, 1 month to 6 years) were included. Varied types of biventricular repair were customized individually. Follow-up was 90.4% complete, and the mean follow-up time was 3.4 ± 3.9 years. There were 17 (4.5%) early deaths and 7 (2.1%) late deaths. Preoperative pulmonary hypertension was the only risk factor for early mortality. Postoperative significant left ventricular outflow tract obstruction was present in 9 survivors. Patients with noncommitted ventricular septal defect had a longer crossclamp time, longer cardiopulmonary bypass time, and higher incidence of postdischarge left ventricular outflow tract obstruction. There were 4 reoperations, all of which were caused by subaortic left ventricular outflow tract obstruction. All of the pressure gradients were decreased to less than 20 mm Hg after the modified Konno procedure with an uneventful postoperative course.

Conclusions: Optimal results of varied types of biventricular repair for double outlet right ventricle have been acquired. Although noncommitted ventricular septal defect is technically difficult, the outcomes of patients are favorable. Late-onset left ventricular outflow tract obstruction is the main reason for reoperation but can be successfully relieved by the modified Konno procedure. (J Thorac Cardiovasc Surg 2014;148:817-24)

Double outlet right ventricle (DORV) is a congenital anomaly in which both the aorta and the pulmonary artery originate from the right ventricle, representing a broad spectrum of anatomic variants and associated malformations.^{1,2} For many years, arguments have persisted on whether the lesion was better defined on the basis of (1) the connections between the arterial trunks and their supporting ventricle or (2) the presence of infundibular musculature supporting exclusively the leaflets of both arterial valves. To avoid misunderstanding, the 50%

override rule was concisely used to define and diagnose DORV in the current study.

The pathophysiology of DORV varies from severe cyanosis to significant volume overload similar to a large ventricular septal defect (VSD) shunt. The optimal surgical approach is tailored to both the anatomic features and their physiologic consequences. A promising approach is highly varied and needs to be customized for individuals.³

Accompanying techniques have improved in the last decade, and a growing proportion of patients are undergoing biventricular repair.⁴ However, the outcomes of anatomic repair for patients with DORV and noncommitted VSD are not as apparent. In addition, late postoperative subaortic obstruction has attracted attention. In the current study, we report our results among 380 patients with DORV presenting for biventricular repair at Fuwai Hospital (Beijing, PR China) and the surgical strategy for late-onset left ventricular outflow tract obstruction (LVOTO).

METHODS

Patients

Patients with DORV who underwent biventricular repair at Fuwai Hospital from January 2005 to December 2012 were included in this retrospective study. Diagnosis of DORV was made if both great arteries originated predominantly from the right ventricle with application of the “50% rule.”⁵ Patients were excluded if DORV was combined with

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Abbreviations and Acronyms

ANOVA	= analysis of variance
DORV	= double outlet right ventricle
IVR	= intraventricular tunnel repair
LVOTO	= left ventricular outflow tract obstruction
RVOT	= right ventricular outflow tract
VSD	= ventricular septal defect

atrioventricular septal defect, heterotaxy syndrome, atrioventricular discordance, or univentricular physiology. According to the location of VSD, patients were divided into group 1 (with noncommitted VSD), group 2 (with subaortic VSD), group 3 (with sub-pulmonic VSD), and group 4 (with doubly committed VSD). This study was approved by the ethics committee at Fuwai Hospital. The hospital gave us approval to waive the need for patient consent for publishing follow-up data on these patients.

Clinical Protocol and Surgical Technique of Biventricular Repair for Double Outlet Right Ventricle

Indication for varied approaches of anatomic repair depended on anatomic features, achieving biventricular repair for these patients. Intraventricular tunnel repair (IVR) to the aorta was performed in patients with VSD-type DORV; IVR to the aorta + right ventricular outflow tract (RVOT) reconstruction was performed in patients with tetralogy-type DORV; IVR + arterial switch operation or double root translocation was performed in patients with transposition-type DORV; and the réparation à l'étage ventriculaire or Rastelli procedure was performed when the pulmonary valve stenosis that was considered could not be enlarged. For DORV and noncommitted VSD, the choice of appropriate procedures was complex. The protocol of Fuwai Hospital is shown in Figure 1, A.

Pulmonary artery banding and modified Blalock-Taussig shunt were used as previous palliative approaches to restrict pulmonary plethora and alleviate cyanosis, respectively. At biventricular repair, all patients underwent operation with standard cardiopulmonary bypass, bicaval cannulation, and moderate hypothermia with cold potassium cardioplegic arrest. Intracardiac baffle was reconstructed by Dacron patch. VSD enlargement and chordae reattachment were selectively performed, substantially reducing the potential for subaortic stenosis and improving the baffle geometry. Both reducing the pulmonary valve regurgitation and avoiding further external conduit replacement were principles when reconstructing the RVOT. The techniques of the arterial switch operation or double root translocation were similar in reports from Morgan Stanley Children's Hospital⁶ and our center.^{7,8}

Clinical Protocol and Surgical Technique for Late-Onset Left Ventricular Outflow Tract Obstruction

At Fuwai Hospital, significant late postoperative LVOTO was defined as a trans-stenosis systolic pressure gradient of 30 mm Hg or greater. Indication for reoperation relieving late-onset LVOTO was stenosis-associating symptoms combined with a trans-stenosis systolic pressure gradient of 50 mm Hg or greater. Stenosis-associating symptoms included angina, syncope, and dyspnea. When the trans-stenosis systolic pressure gradient was greater than 75 mm Hg, surgical intervention was aggressively indicated regardless of symptoms.

The modified Konno was established as the procedure of choice to relieve postdischarge subaortic LVOTO after biventricular repair for DORV. Cardiopulmonary bypass techniques included continuous flow with bicaval cannulation, blood prime, and moderate hypothermia. Myocardial protection was achieved with antegrade single-dose blood cardioplegia. Previous intracardiac baffle, partial ventricular septal, and

subaortic conus were carefully resected after right ventriculotomy. The incised area was limited to the upper part of the trabecular septum, and thus injury to the conduction system and major septal coronary arteries could be avoided. The ventricular septal was enlarged, and the new intracardiac baffle was reconstructed.

Data Collection and Definition

Patient demographics and clinical data were obtained from our local database. Ventricle function, size, and valve stenosis/regurgitation were assessed by echocardiography. During follow-up, patients were contacted by telephone or direct interview in our outpatient clinic every 3 to 6 months. A single cardiologist reviewed all previous echocardiograms and performed independent measurements. Pulmonary arterial hypertension was defined as mean pulmonary artery pressure more than 25 mm Hg. The severity of the valvular regurgitation was graded according to guidelines published by the American Society of Echocardiography.⁹ Valvular regurgitation was considered significant when documented as moderate or severe, and ventricular dysfunction was defined as an ejection fraction less than 50%. In-hospital mortality was defined as both 30-day mortality and death any time after operation but before discharge. Postdischarge mortality was defined as death after 30 days or after discharge if the length of hospital stay was more than 30 days. Reoperation included only reoperations on the heart and excluded secondary closure of the sternum and revision for bleeding or mediastinitis.

Statistical Analysis

Continuous variables were presented as mean \pm standard deviation or median with minimum and maximum range, and categorical variables were presented as percentage. Comparisons of variables were made using the Student *t* test, analysis of variance (ANOVA), chi-square test, or Fisher exact test. Bonferroni test was used in pairwise comparison. Time to death and time to postoperative LVOTO are displayed by Kaplan-Meier curves. Logistic regression (forward) was performed as multivariate analysis to investigate risk factors for early death, late death, and postoperative LVOTO. All entered variables were selected on the basis of clinical experience, univariate analysis, and previously published data. To find risk factors for early and late deaths, variables that entered the model included age less than 1 year at repair, preoperative pulmonary arterial hypertension, noncommitted VSD, aortic arch obstruction, previous palliation, pulmonary stenosis, and great arteries relationship. To find risk factors for postdischarge LVOTO, variables entered into the model included age less than 1 year at repair, noncommitted VSD, subaortic conus, pulmonary stenosis, and great arteries relationship. The level of significance was set at an alpha level of 0.05 or less. Analysis was conducted using SPSS version 17.0 (IBM-SPSS Inc, Armonk, NY) for Windows.

RESULTS

Patients' Characteristics and Anatomic Features

From January 2005 to December 2012, 380 consecutive patients with a mean age of 1.9 ± 2.1 years (range, 1 month to 6 years) underwent biventricular repair of DORV at Fuwai Hospital. The mean weight at biventricular repair was 12.2 ± 5.9 kg (range, 5.5-16 kg).

In our local database (not limited to 380 biventricular repairs), 25 palliative procedures were required in 25 patients (pulmonary artery banding and modified Blalock-Taussig shunt). There was no mortality in these patients, and 20 of them had undergone biventricular repair, who were included in this analysis. A total of 20 of 380 patients (5.3%) had a palliative procedure preceding complete repair. All of these patients required only 1 palliative

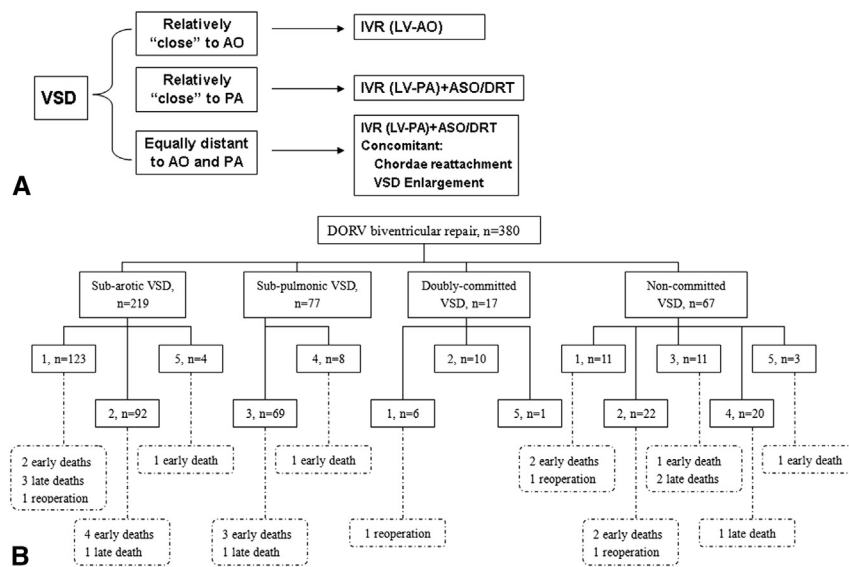


FIGURE 1. A, Protocol for DORV with noncommitted VSD. B, Surgical procedures, death, and reoperations. 1 = IVR to the aorta; 2 = IVR to the aorta + RVOT enlargement; 3 = IVR to the pulmonary artery + arterial switch operation; 4 = IVR to the pulmonary artery + double root translocation; 5 = IVR to the aorta + Rastelli/réparation à l'étage ventriculaire. AO, Aorta; ASO, arterial switch operation; DORV, double outlet right ventricle; DRT, double root translocation; IVR, intraventricular tunnel repair; LV, left ventricle; PA, pulmonary artery; VSD, ventricular septal defect.

procedure. Data on VSD position, great arteries, associated anomalies, and previous palliative procedures are shown in Table 1.

As shown in Table 1, there are 67 patients (17.6%) in group 1, 219 patients (57.6%) in group 2, 77 patients (20.2%) in group 3, and 17 patients (4.5%) in group 4. Compared with patients in all other groups, patients in group 1 had more subaortic conus (chi-square test: $P < .001$; pairwise comparison: vs group 2, $P < .001$; vs group 3, $P < .001$; vs group 4, $P = .01$). Moreover, there were more pulmonary stenoses in group 1 than in groups 2 and 3 (chi-square test: $P < .001$; pairwise comparison: vs group 2, $P = .011$; vs group 3, $P < .001$; vs group 4, $P = .848$).

Surgical Procedures

A total of 380 biventricular repair approaches were performed, including (1) IVR to the aorta ($n = 140$, 36.9%); (2) IVR to the aorta + RVOT enlargement ($n = 124$, 32.6%); (3) IVR to the pulmonary artery + arterial switch operation ($n = 80$, 22.1%); (4) IVR to the pulmonary artery + double root translocation ($n = 28$, 7.4%); and (5) IVR to the aorta + Rastelli/réparation à l'étage ventriculaire ($n = 8$, 2.2%). Data are shown in Figure 1, B. Concomitant procedures included VSD enlargement in 23 patients (6.1%), chordae reattachment in 7 patients (1.9%), atrioventricular valvuloplasty in 12 patients (3.2%), and aortic sinus plasty in 3 patients (0.8%).

Early Results

Both crossclamp time (ANOVA: $P = .044$; pairwise: vs group 2, $P < .001$, vs group 3, $P = .008$; vs group 4,

$P = .013$) and cardiopulmonary bypass time (ANOVA: $P = .029$; vs group 2, $P < .001$; vs group 3, $P = .010$; vs group 4, $P = .010$) were longer in group 1 than in all other groups. However, ventilation time (ANOVA: $P = .322$) and intensive care unit stay (ANOVA: $P = .556$) were similar between groups.

Among all 17 (4.4%) in-hospital deaths, 6 were caused by pulmonary hypertension crisis, 6 were caused by low cardiac output syndrome, 4 were caused by pulmonary infection, and 1 was caused by malignant arrhythmia. Two patients required implantation of a permanent pacemaker for surgically induced complete heart block. Preoperative pulmonary artery hypertension (odds ratio, 5.2; $P = .020$) was identified as the only risk factor for early deaths in multivariate analysis.

Follow-up Outcomes

Follow-up information was available for 341 survivors (90.4%), and the mean follow-up time was 3.4 ± 3.9 years (range, 8 months to 9 years). According to up-to-date follow-up records, there were 7 (2.1%) late deaths. The causes were cardiac failure in 2 patients, pulmonary hypertension crisis in 2 patients, sudden death in 1 patient, and noncardiac in 2 patients. All late deaths occurred within 1 year after biventricular repair. In multivariate analysis, there was no variable identified as an independent risk factor for postdischarge deaths. Figure 2, A displays the overall estimated survival, including operative mortality, which was 94.4%, 93.5%, and 93.5% at 6 months, 1 year, and 5 years, respectively. As Table 1 shows, there were trends that

TABLE 1. Patient characteristics and surgical results

Variables	Total cohort (n = 380)	ncVSD (n = 67)	Subaortic VSD (n = 219)	Subpulmonic VSD (n = 77)	Doubly committed VSD (n = 17)
Patient characteristics					
Mean age at BVR (y)	1.9 ± 2.1	2.4 ± 1.6	1.2 ± 1.1	0.5 ± 0.7	1.5 ± 1.6
Mean weight at BVR (kg)	12.2 ± 5.9	15.3 ± 8.0	9.6 ± 5.0	5.9 ± 7.1	12.2 ± 8.3
Great arteries					
Normal relation	193 (50.8%)	32 (47.8%)	130 (59.3%)	20 (26.0%)	11 (64.7%)
Side by side	59 (15.5%)	16 (23.9%)	12 (5.5%)	30 (39.0%)	1 (5.9%)
Anterior aorta	128 (33.7%)	19 (28.5%)	77 (35.2%)	27 (35.1%)	5 (29.4%)
Subaortic conus	197 (51.8%)	52 (77.6%)	117 (53.4%)	22 (28.6%)	6 (35.3%)
Pulmonary stenosis	166 (43.7%)	45 (67.2%)	96 (43.9%)	14 (18.2%)	11 (64.7%)
Coronary anomalies	69 (18.2%)	20 (29.9%)	25 (11.5%)	21 (27.3%)	3 (17.6%)
Arch obstruction	12 (2.9%)	1 (1.5%)	3 (1.4%)	7 (9.1%)	1 (5.9%)
Pulmonary arterial hypertension	76 (20.0%)	12 (17.9%)	43 (19.6%)	16 (20.8%)	5 (29.4%)
Previous palliative procedure					
Pulmonary artery banding	9 (2.4%)	6 (9.0%)	2 (0.9%)	1 (1.3%)	0
Modified B-T shunt	11 (2.9%)	2 (3.0%)	6 (2.7%)	3 (3.9%)	0
Outcomes					
CPB time (min)	148 ± 55	170 ± 50	137 ± 61	144 ± 45	140 ± 49
Crossclamp time (min)	62 ± 30	97 ± 31	56 ± 38	69 ± 50	62 ± 43
Ventilation time (h)	32.5 ± 25.8	44.7 ± 28.3	30.2 ± 28.5	28.0 ± 21.4	31.7 ± 22.0
ICU stay (d)	6.2 ± 5.0	6.8 ± 5.8	5.6 ± 6.3	5.2 ± 4.7	6.0 ± 4.1
In-hospital mortality	17 (4.5%)	6 (8.9%)	7 (3.2%)	4 (5.2%)	0
Late mortality	7 (2.1%)	3 (5.5%)	3 (1.5%)	1 (1.5%)	0
Follow-up completeness	341 (90.4%)	55 (88.8%)	203 (95.8%)	68 (93.1%)	15 (88.2%)
NYHA class III-IV	1 (0.3%)	0	1 (0.5%)	0	0
LVEF (%)	66 ± 14	63 ± 10	62 ± 13	67 ± 9	67 ± 10
Early LVOTO	6 (1.7%)	4 (6.6%)	2 (0.9%)	0	0
Late-onset LVOTO	9 (2.7%)	6 (11.5%)	2 (1.0%)	1 (6.7%)	0

B-T, Blalock-Taussig; BVR, biventricular repair; CPB, cardiopulmonary bypass; ICU, intensive care unit; LVEF, left ventricular ejection fraction; LVOTO, left ventricular outflow tract obstruction; ncVSD, noncommitted ventricular septal defect; NYHA, New York Heart Association; VSD, ventricular septal defect.

in-hospital mortality and postdischarge mortality were higher in group 1 than in other groups. However, neither of them reached statistical significance (chi-square test: in-hospital mortality, $P = .181$; postdischarge mortality, $P = .298$). For patients in group 1, estimated overall survival was 89.5%, 86.2%, and 86.2% at 6 months, 1 year, and 5 years, respectively. For patients in group 2, estimated overall survival was 96.3% at 6 months and 95.2% at 1 year and 5 years. For patients in group 3, estimated overall survival was 94.8%, 94.8%, and 94.8% at 6 months, 1 year, and 5 years, respectively. For patients in group 4, estimated overall survival was 100.0% at 6 months, 1 year, and 5 years. There were no statistical differences among the survival curves in these 4 groups ($P = .891$).

No coronary lesion, significant neo-aortic regurgitation, or RVOT obstruction was noted at the latest echocardiography in patients who underwent the arterial switch operation. In patients who underwent RVOT reconstruction, pulmonary valve regurgitation was found in only 1 patient, and he is awaiting reintervention. At the latest follow-up, mean trans-stenosis pressure gradient of the RVOT was 26.8 ± 15.5 mm Hg. There was no statistical significance in New York Heart Association functional

class III to IV (chi-square test, $P = 1.000$) and mean left ventricular ejection fraction (ANOVA: $P = .955$) between these groups.

Postoperative Left Ventricular Outflow Tract Obstruction

As the result of unfavorable intracardiac tunnel geometry, early baffle revision was required immediately after biventricular repair in 6 patients.

There were 9 (2.7%) significant postdischarge LVOTOs. The mean duration from biventricular repair to postdischarge LVOTO occurrence was 55.6 ± 23.3 months, and the mean pressure gradient was 73.8 ± 40.2 mm Hg. All stenoses were at the level of baffle-patch, septal, or muscular structures, which was confirmed by transthoracic echocardiography.

Probability of freedom from postoperative LVOTO (both pre- and postdischarge) was 98.4% at 1 year, 96.3% at 3 years, and 92.8% at 10 years in the total population. The characteristics of all these 15 patients are shown in Table 2. In multivariate analysis, age at biventricular repair of less than 1 year, noncommitted VSD, and subaortic conus were risk factors for postoperative LVOTO.

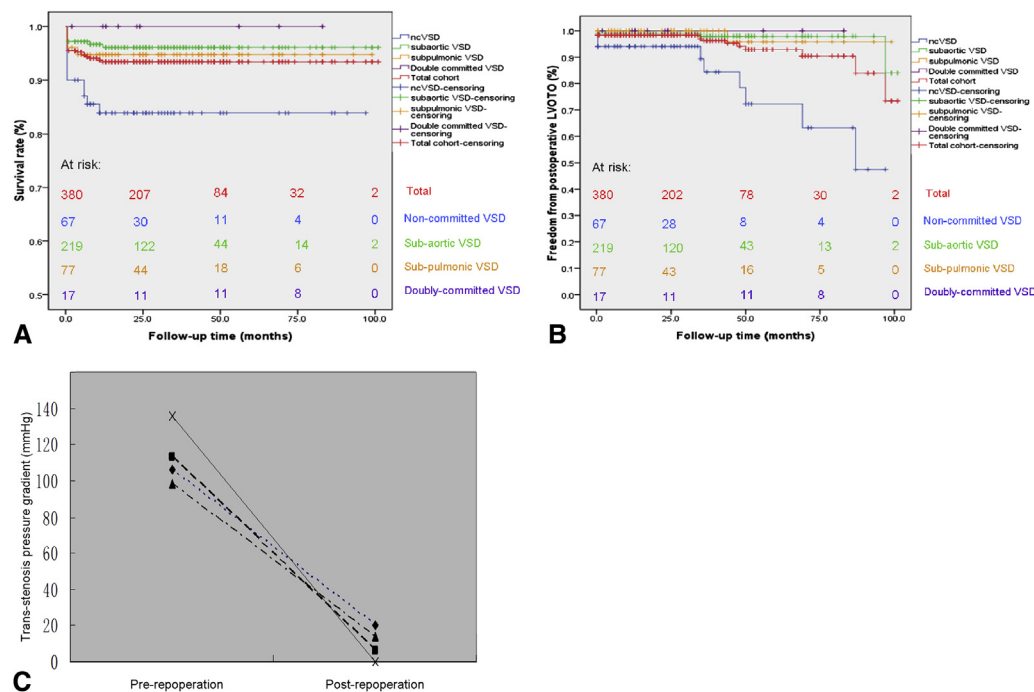


FIGURE 2. A, Kaplan–Meier curves for mortality (B) Kaplan–Meier curves for total postoperative LVOTO. C, Decrease of trans-stenosis pressure gradient after reoperation. *LVOTO*, Left ventricular outflow tract obstruction; *ncVSD*, noncommitted ventricular septal defect; *VSD*, ventricular septal defect.

There were more early LVOTOs (chi-square test: $P = .015$; pairwise comparison: vs group 2, $P = .011$) and postdischarge LVOTOs (chi-square test: $P < .001$; pairwise comparison: vs group 2, $P < .001$) in group 1 than in

group 2. For patients in group 1, the probability of freedom from postoperative LVOTO was 94.0%, 84.4%, and 72.3% at 1, 3, and 5 years, respectively. For patients in group 2, freedom from postoperative LVOTO was 99.1% at 1 year

TABLE 2. Characteristics of patients with postdischarge left ventricular outflow tract obstruction

	Age at repair (mo)	Systolic pressure gradient (mm Hg)	Duration from biventricular to LVOTO (mo)	VSD position	Subaortic conus	Pulmonary stenosis	Great arteries	Procedure
Early LVOTO								
1	20	43	Immediately after BVR	Noncommitted	+	–	Anterior aorta	IVR
2	6	66	Immediately after BVR	Noncommitted	+	+	Normal	IVR
3	9	50	Immediately after BVR	Noncommitted	+	–	Anterior aorta	IVR
4	10	75	Immediately after BVR	Subaortic	–	–	Anterior aorta	IVR
5	12	55	Immediately after BVR	Subaortic	+	+	Normal	IVR + RVOTE
6	5	30	Immediately after BVR	Noncommitted	+	+	Anterior aorta	IVR
Late-onset LVOTO								
7	11	106	44	Doubly committed	+	–	Normal	IVR
8	11	98	35	Noncommitted	+	+	Anterior aorta	IVR + RVOTE
9	7	39	48	Noncommitted	+	–	Anterior aorta	IVR
10	7	33	35	Subaortic	+	–	Anterior aorta	IVR
11	5	43	50	Noncommitted	+	–	Anterior aorta	IVR
12	6	113	69	Noncommitted	+	–	Anterior aorta	IVR
13	12	35	87	Noncommitted	+	+	Anterior aorta	IVR
14	24	136	97	Subaortic	+	+	Anterior aorta	IVR
15	4	67	36	Noncommitted	+	–	Anterior aorta	IVR

BVR, Biventricular repair; *IVR*, intraventricular tunnel repair; *LVOTO*, left ventricular outflow tract obstruction; *RVOTE*, right ventricular outflow tract enlargement; *VSD*, ventricular septal defect.

TABLE 3. Characteristics of patients who underwent modified Konno procedure

	Age at reoperation (y)	Duration from BVR to reoperation (mo)	LVEF before reoperation (%)	Pre-reoperative systolic pressure gradient (mm Hg)	Post-reoperative systolic pressure gradient (mm Hg)
1	4.8	48	66	106	19
2	4.9	39	70	113	8
3	6.6	90	63	98	14
4	10.0	100	79	136	3

BVR, Biventricular repair; LVEF, left ventricular ejection fraction.

and 98.0% at 3 and 5 years. Freedom from postoperative LVOTO at 5 years was 95.8% and 100% in groups 3 and 4, respectively.

Among the 9 patients who had postdischarge LVOTO, 4 underwent 4 reoperations (modified Konno procedure). The characteristics of these 4 patients are shown in Table 3. The mean crossclamp time was 62 ± 33 minutes, mean cardiopulmonary bypass time was 103 ± 47 minutes, and mean intensive care unit stay time was 5.3 ± 4.4 days. There was no early mortality in patients undergoing these reoperations. Moreover, mean left ventricular ejection fraction was $65.5\% \pm 9.3\%$. No conduction blockade was found. All the pressure gradients were reduced to less than 20 mm Hg (Figure 2, C). During a relatively short follow-up period (1.4 ± 1.2 years; range, 5 months to 2.7 years), there was no late mortality and all pressure gradients remained less than 20 mm Hg.

DISCUSSION

In a period of only 7 years, 380 patients with DORV have undergone biventricular repair at Fuwai Hospital. This study analyzing data of this large cohort confirms that a biventricular repair can be achieved with favorable outcomes in the majority of patients with DORV.

As shown in Figure 1 and stated in the “Methods” section, varied surgical strategies were used for biventricular repair. Evaluation of anatomic factors influencing the surgical decision (eg, VSD location, presence of conus, right ventricular tract obstruction, atrioventricular valve, and balance of ventricles) is essential to obtain favorable outcomes.

Among these 380 patients with DORV who underwent biventricular repair, 17 early deaths and 7 late deaths were documented. Consistent with Brown and colleagues¹⁰ precise conclusion, both in-hospital mortality and late mortality were acceptable. Five-year overall survival of 93.5% in this study is similar to that reported by others.^{11,12} Bradley and colleagues¹³ reported 56% 15-year survival, which was lower. However, they selected patients with more complex anatomy and focused on outcomes after biventricular repair or univentricular palliation.

In our experience, we pursued primary neonatal repair in the majority of patients with DORV. However, we prefer palliation followed by biventricular repair if the VSD is

truly remote. It may be easier to establish an unobstructed intracardiac baffle when accomplishing biventricular correction beyond the age of 3 months. The mean age at biventricular repair was 1.9 ± 2.1 years in our cohort. In developing countries, late referral of patients is common and accompanied pulmonary arterial hypertension is a medical issue of concern.¹⁴ In the current study, a notable amount of early deaths were directly caused by pulmonary arterial hypertension crisis. Moreover, the presence of preoperative pulmonary arterial hypertension was identified as the only independent risk factor for early deaths in multivariate analysis. Thus, postoperative oral drugs to reduce pulmonary arterial pressure are strongly indicated in patients who have preoperative pulmonary arterial hypertension. However, inadequate follow-up in detecting and treating pulmonary hypertension has been common in China, and the compliance with oral drug therapy remains unsatisfactory. These are the reasons for the 2 late deaths caused by late pulmonary arterial crisis. Unlike in previous studies,^{10-13,15} in our cohort, all late deaths occurred within 1 year after biventricular repair, representing a stable and safe long-term postoperative course.

DORV with noncommitted VSD represents the most extreme form of DORV, raising surgical difficulties for biventricular repair. The definition of DORV with noncommitted VSD includes (1) a VSD distant (greater than aortic diameter) from both arterial valves; (2) both great vessels arising fully from the right ventricle; and (3) a double conus.^{16,17} As Brown and colleagues¹⁰ stated, among patients with noncomplex forms of DORV and noncommitted VSD, biventricular repair is still possible in the majority of patients with acceptable results. In the current study, although both crossclamp and cardiopulmonary bypass times were longer in patients with noncommitted VSD, ventilation time, intensive care unit stay, mortality, and cardiac function were statistically similar between these 2 groups. All of these findings show that the results of patients with DORV and noncommitted VSD are favorable, although these conditions are technically more difficult (represented by prolonged crossclamp time and cardiopulmonary bypass time).

There were 9 (2.7%) late-onset LVOTOs in the total cohort. Except for appropriate surgical management, another potential reason for our low incidence of late-onset LVOTO is the relatively short follow-up period. As

Figure 2, B, and Table 2 show, all durations from biventricular repair to the presence of late-onset LVOTO were approximately more than 3 years, suggesting that its development is somatic outgrowth relating. Moreover, the finding that younger age at biventricular repair was a risk factor for late-onset LVOTO confirms this speculation. We realized that our material for intracardiac baffle was a Dacron patch, which is unable to grow. Applying other baffle materials may decrease the incidence of late-onset LVOTO.

We are in agreement with Belli and colleagues¹⁸ that LVOTO after “IVR to the aorta” repair of DORV develops at 3 levels: aortic annulus, baffle patch/muscular structures, and VSD. Rychik and coworkers¹⁹ analyzed the preoperative and postoperative echocardiograms of 24 patients who underwent IVR for DORV and showed that the left ventricle undergoes geometric changes in which the VSD is used as the new left ventricular outflow pathway. Instead of the aortic annulus and tunnel entry stenosis, the bulge of the underlying septal or subaortic conal muscle was the main cause of obstruction in all 9 patients who had late-onset LVOTO.

Postdischarge LVOTO is a significant complication of patients with DORV and noncommitted VSD, which is confirmed by our results. Compared with group 2, group 1 had statistically more postdischarge LVOTO. Although statistically insignificant, there is a trend that more LVOTO developed in group 1 than in groups 3 or 4. The small sample size may be the reason for our failure in detecting these statistical differences. In the past, IVR of the VSD to the aorta was the operation of reference for DORV with noncommitted VSD. With this technique, the incidence of postdischarge subaortic obstruction was evaluated at 30% by Belli and colleagues.^{18,20} In our cohort, only 6 patients (11.5%) in group A developed significant postdischarge LVOTO during follow-up, probably benefiting from the use of the IVR to the pulmonary artery combined with the arterial switch (Figure 1 shows the surgical protocol). The short length of the tunnel is the key advantage of this technique because the longer the tunnel is, the greater the risk of subaortic obstruction.

In our experience, the principle of reoperation for subaortic LVOTO is aimed not only at relieving the obstruction but also at streamlining the left ventricular outflow tract by removal of protrusions located at the subaortic level. Rocchini and colleagues²¹ proposed that kinking and shrinkage of the baffle with time are possible mechanisms leading to postdischarge LVOTO. By VSD enlargement and tunnel reestablishment, all of the pressure gradients were reduced to less than 20 mm Hg in our patients who underwent reoperation, which was similar to the result reported by Belli and colleagues.¹⁸ Moreover, the approach we described seems technically feasible with relatively short crossclamp and cardiopulmonary times. However, the follow-up duration

is not long enough, and further reobstruction requires careful evaluation.

Study Limitations

Limitations of this study include its single institution and retrospective nature. The number of patients lost to follow-up may have influenced the interpretation of the results. Objective assessments of functional capacity (eg, cardiopulmonary exercise testing) and further follow-up are needed in future studies.

CONCLUSIONS

Optimal results of varied types of biventricular repair for DORV have been acquired. Although noncommitted VSD is technically difficult, the outcomes of patients are favorable. Late-onset LVOTO is the main reason for reoperation but can be successfully relieved by the modified Konno procedure.

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